



Focal Treatment for Genetic Disorders (A Fairy Tale or a True Story?)

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Genetic is widely involved in pathophysiology of many diseases. However, certain gene abnormalities could cause cardiac disorders called *monogenic disorders* (1).

Since the genetic information and hence abnormal gene is present in all cells, treatments are focused on symptoms. Nowadays, gene transfer is an uprising option (2).

Although all cells carry a mutant gene, the major effect of the disease is localized to a group of cells in some patients.

Brugada syndrome results from a sodium channel mutation that causes ST elevation in right precordial leads and may lead to sudden ventricular fibrillation, especially in young males (3). This main electrocardiographic disturbance originates from RV epicardium (4). Therefore, some interventionists try to treat the disease by ablation of this area (5). Although implanting a defibrillator is yet the standard recommendation, it does not cure the disease (6).

Thus, ablation of a small area of epicardium which offers a definitive cure also seems appealing.

Another example of applying this treatment for a genetic disorder is in catecholaminergic polymorphic ventricular tachycardia (7).

This syndrome is caused by a mutation in ryanodine receptor and manifests as polymorphic ventricular tachycardia during exercise or emotion in young adults (8). One study showed that ablation of Premature Ventricular Contraction (PVC) completely treated a patient (7). Although myocardial contraction duration is prolonged in all segments (9) in the patients with long QT syndrome, there is a segment with most prolonged duration in any patient (10). Ablation of PVC on this segment may cure the Ventricular Fibrillation (VF) in this syndrome and prevent sudden cardiac death (11).

Another lethal and seemingly incurable syndrome of sudden cardiac death is idiopathic VF. As sounded from nomenclature, the pathophysiology is somewhat unclear and, consequently, the treatment is difficult and only limited to secondary prevention (12). However, ablation of PVC starting the VF looks effective and possible (11). Another recently noticed disease of sudden cardiac disease is early repolarization, especially if it is localized to the inferolateral area (13). Michel Haïssaguerre has described few cases of ablation for this syndrome (14).

In conclusion, genetic problems play a significant role in cardiac diseases. They have a widespread nature and present in all body cells; hence, their handling is quite difficult.

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Palliation therapy and genetic engineering have led to some improvement in management. Yet, many shortages are still present.

Of course, simple solutions might be uprising for these hereditary diseases. The clues of this local approach can be found in the case reports mentioned in this article. Of course, more studies and trials are required to be conducted on this approach.

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